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Ruptured Lenticulostriate Artery Aneurysm Associated with Moyamoya Disease: A Case Report and Literature Review.

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Abstract

INTRODUCTION: Clinically, ruptured lenticulostriate artery (LSA) aneurysm associated with moyamoya disease (MMD) is rare but represents a potential hemorrhagic risk. Its optimal management remains unknown.
CASE DESCRIPTION: A 66-year-old woman developed a left basal ganglia hemorrhage with intraventricular extension secondary to an MMD-associated distal LSA aneurysm that was subsequently treated with endovascular embolization. In this report, we review all previous cases of ruptured LSA aneurysms related to MMD.
CONCLUSION: LSA aneurysm rupture should be considered in the setting of hemorrhagic MMD, especially in combination with basal ganglia hematoma. Proximal and distal LSA aneurysms appear to have different types of hemorrhage. This case highlights that management of such aneurysms should be individualized based on the balance of benefits and risks.

CASE REPORT

INTRODUCTION

Lenticulostriate arteries (LSAs) arise from the proximal portions of the anterior and middle cerebral arteries and are traditionally divided into medial and lateral groups, supplying the basal ganglia, most of the internal capsule, and the anterior commissure. In the setting of moyamoya disease (MMD), the LSAs may serve as a collateral supply to the anterior circulation, making them susceptible to LSA aneurysm formation due to long-term hemodynamic stress (Chalouhi *et al.* 2013; Fu *et al.* 2022). The presence of such an aneurysm may pose a potential hemorrhagic risk, but there is no consensus on the best way to treat it (Sung *et al.* 2011; Feng *et al.* 2024).

We herein present a case of a 66-year-old woman with a ruptured LSA aneurysm associated with MMD that was subsequently treated by endovascular embolization. Moreover, the relevant literature is reviewed and the clinical features, as well as the treatment strategy, are also discussed.

CASE PRESENTATION

A 66-year-old woman with a history of hypertension presented with a sudden onset of severe headache, nausea and right-sided weakness. Neurological examination showed a 3/5 weakness in the right upper and lower extremities. Initial computed tomography of the head revealed a left basal ganglia hematoma with intraventricular extension (Figure 1A). Magnetic resonance angiography showed complete occlusion of the bilateral internal carotid arteries, consistent with the diagnosis of MMD (Figure 1B). Catheter-based angiography demonstrated definitive MMD and an aneurysm arising from the distal segment of the left LSA (Figure 2A-B). The aneurysm was thought to be responsible for the hemorrhage and treatment was indicated.

Endovascular embolization was scheduled as the first-line treatment. Briefly, under general anesthesia, a 6-French guiding catheter was positioned in the left internal carotid artery through which a Marathon-10 microcatheter was advanced over a Synchro-10 microguidewire and navigated into the left LSA proximal to

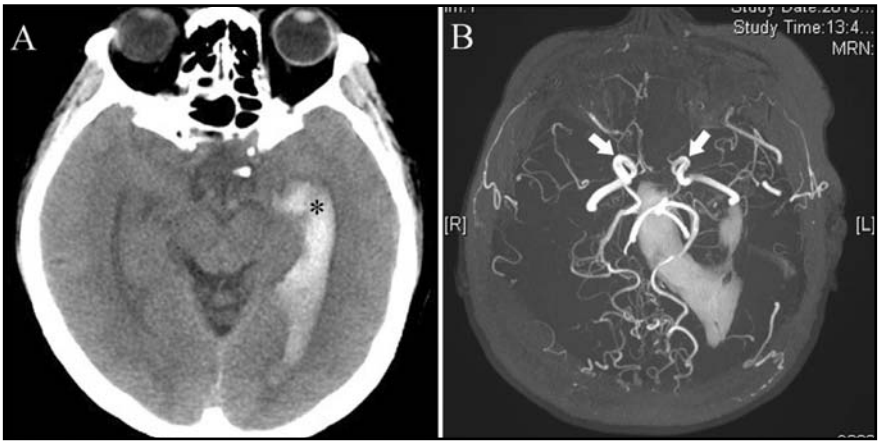

Fig. 1. (A) Brain computed tomography showed a left basal ganglia hemorrhage (asterisk) extending into the ventricular system. (B) Magnetic resonance angiography showed complete occlusion at the terminal portion of both internal carotid arteries (arrows), indicating the diagnosis of moyamoya disease.

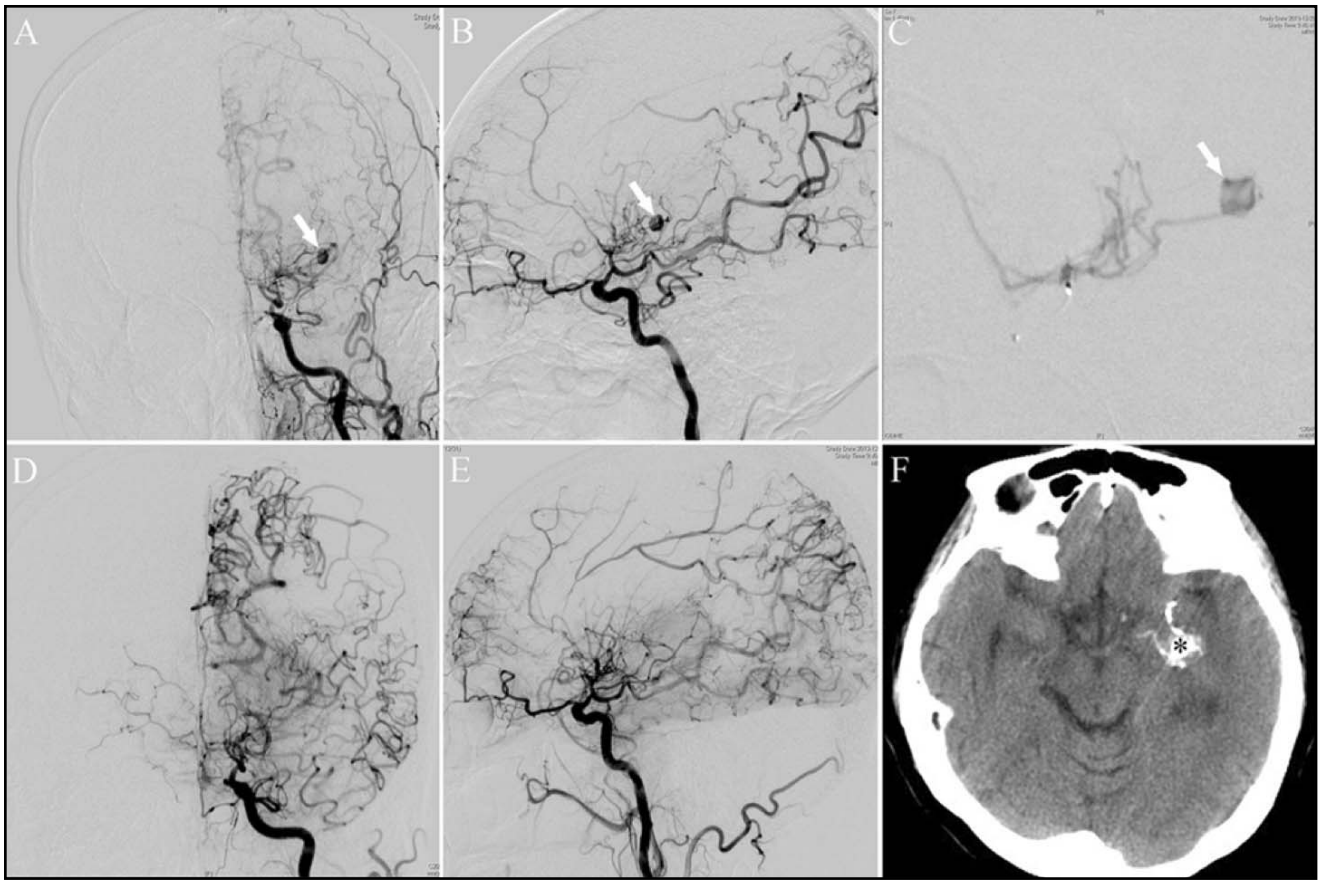

Fig. 2. (A, B) Catheter-based angiograms revealed a typical collateral vascular network of moyamoya disease and an incidental flow aneurysm (arrow) distal on the left lenticulostriate artery (LSA). (C) Superselective angiography before glue injection confirmed the distal LSA aneurysm (arrow). (D, E) Postembolization angiograms showed complete obliteration of the aneurysm. (F) Postoperative brain computed tomography showed the liquid embolic agent filling in the LSA and aneurysm (asterisk).

the aneurysm sac. A 20% mixture of Glubran 2 cyanoacrylate glue and lipiodol was then slowly injected into the LSA up to the aneurysm (Figure 2C). The immediate angiogram revealed no aneurysmal filling (Figure 2D-E). Her neurological status remained stable postoperatively. Repeat head computed tomography showed the aneurysm and target branch filled with liquid embolic agent and no infarction in the left LSA territory (Figure 2F). At the 10-year follow-up, the patient had a complete recovery.

DISCUSSION

Ruptured LSA aneurysms associated with MMD are relatively rare; to the best of our knowledge, only 23 cases with sufficient individual information have been reported in the English literature to date, including the present case (Ahn *et al.* 2007; Ando *et al.* 2023; Bechan & van Rooij, 2014; Byeon *et al.* 2022; Chalouhi *et al.* 2013; Fu *et al.* 2022; Grabel *et al.* 1989; Gandhi *et al.* 2008; Hwang *et al.* 2014; Hashio *et al.* 2024; Larrazabal *et al.* 2001; Liu *et al.* 2016; Ni *et al.* 2018; Oka *et al.* 1991; Sakai *et al.* 2005; Sung *et al.* 2011; Zhou *et al.* 2024). General characteristics of these patients are summarized in Table 1.

Regarding ethnicity, 7 patients (30.4%) were identified as Caucasian, 7 (30.4%) as Chinese, 5 (21.7%) as Korean, and 4 (17.5%) as Japanese. The mean age was 46.6 years, ranging from 10 to 73 years. There were 12 women and 11 men, with a female-to-male ratio of 1.1:1. Initial symptom information was available for 18 cases, and the most common presentation was impaired consciousness (8/18, 44.4%) and headache (8/18, 44.4%), followed by hemiparesis (4/18, 22.2%).

In this study, aneurysm location was dichotomized as proximal or distal. As shown in Table 1, 4 aneurysms (4/23, 17.4%) were located in the proximal LSA and 19 (19/23, 82.6%) were located in the distal LSA. Of these 23 cases, the hemorrhage pattern included intracerebral hemorrhage (ICH), intraventricular hemorrhage (IVH), subarachnoid hemorrhage (SAH), or a combination of these. Notably, the type of bleeding differed between proximal and distal aneurysms. Proximal LSA aneurysms tended to manifest as ICH+SAH (2/4, 50.0%), followed by isolated SAH (1/4, 25.0%), and a combination of all three (1/4, 25.0%). In contrast, distal LSA aneurysms were more likely to present with ICH+IVH (8/19, 42.1%), followed by isolated ICH (6/19, 31.6%), isolated IVH (3/19, 15.8%), and ICH+SAH (2/19, 10.5%). As in this case, the most common location of ICH was the basal ganglia (17/19, 89.5%) (Ahn *et al.* 2007; Ando *et al.* 2023; Bechan & van Rooij, 2014; Chalouhi *et al.* 2013; Fu *et al.* 2022; Grabel *et al.* 1989; Gandhi *et al.* 2008; Hwang *et al.* 2014; Hashio *et al.* 2024; Larrazabal *et al.* 2001; Ni *et al.* 2018; Sakai *et al.* 2005; Sung *et al.* 2011).

In MMD patients, LSA aneurysms may predispose to hemorrhagic stroke if left untreated. However, such lesions present diagnostic and therapeutic challenges. First, their size is generally too small to be easily missed by routine neuroimaging, and catheter-based angiography has been widely used to identify these aneurysms in previous studies. Second, their deep location and the fragility of the vessel wall make both microsurgical and endovascular surgery difficult (Byeon *et al.* 2022; Feng *et al.* 2024). Third, there is no consensus on the optimal therapy, and their treatment varies, including direct intervention (endovascular embolization, surgical clipping or excision), indirect revascularization surgery, and conservative observation. Among the 23 cases, 11 (47.8%) received endovascular embolization (LSA sacrifice, 10; no LSA sacrifice, 1), 7 (30.4%) underwent direct open surgery (excision, 6; neck clipping, 1), 3 (13.0%) were managed conservatively, and 2 (8.7%) were treated with revascularization bypass. It is noteworthy that aneurysm embolization with sacrifice of the LSA was not associated with postoperative infarction complications as shown in our case (Ando *et al.* 2023; Byeon *et al.* 2022; Chalouhi *et al.* 2013; Hwang *et al.* 2014; Larrazabal *et al.* 2001; Zhou *et al.* 2024).

Fortunately, the overall outcome of ruptured LSA aneurysms in MMD patients appeared to be favorable (20/23, 87.0%). Good outcomes were achieved in 100% (2/2), 90.9% (10/11), 85.7% (6/7), and 66.7% (2/3) of cases treated with revascularization, embolization, direct surgery, and observation, respectively. Further studies with large numbers of patients are warranted to determine the treatment efficacy of the above approaches.

CONCLUSION

LSA aneurysm rupture should be considered in patients with hemorrhagic MMD, especially in the presence of basal ganglia hemotoma, and catheter-based angiography is required when possible. Proximal LSA aneurysms tend to cause ICH+SAH, whereas distal LSA aneurysms cause ICH+IVH. Although no consensus has been reached on the optimal management of MMD-associated ruptured LSA aneurysms, it seems that treatment should be individualized based on weighing the benefits and risks.

ETHICS STATEMENT

Informed consent was obtained from the patient.

CONFLICT OF INTEREST

The authors declare that they have no conflict of interest associated with this manuscript.

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Not applicable.

Tab. 1. Previously reported cases of ruptured LSA aneurysms associated with MMD

Year	Authors	Age/sex	Presentation	Aneurysm location	Hemorrhagic type	Treatment	Outcome
1989	Grabel *et al.*	60/M	Comatose, right hemiparesis	Distal LSA	ICH (BG)	Observation → spontaneous near-complete regression	Good
1991	Oka *et al.*	32/M	Headache, neck stiffness	Proximal LSA	SAH	Observation	Good
2001	Larrazabal *et al.*	57/F	Comatose	Distal LSA	ICH (BG) + IVH	Embolization with the LSA sacrifice	Poor
2005	Sakai *et al.*	61/M	Comatose	Distal LSA	ICH (BG)	Surgical clipping	Good
2007	Ahn *et al.*	49/M	Headache, vomiting	Distal LSA	ICH (BG) + IVH	Observation → rebleeding → surgical excision	Good
2008	Gandhi *et al.*	59/M	Comatose, left hemiparesis	Proximal LSA	ICH (BG) + IVH + SAH	Surgical excision	Good
		37/F	Seizures	Proximal LSA	ICH (BG) + SAH	Surgical excision	Poor
		31/F	Comatose	Proximal LSA	ICH (BG) + SAH	Surgical excision	Good
2011	Sung *et al.*	66/M	Headache	Distal LSA	ICH (BG) + IVH	Surgical excision	Good
2013	Chalouhi *et al.*	49/M	Headache, right hemiparesis, motor aphasia	Distal LSA	ICH (BG)	Embolization with the LSA sacrifice	Good
2014	Bechan and van Rooij	24/F	Headache, nausea, vomiting	Distal LSA (RAH*)	ICH (BG) + SAH	Proximal RAH coiling	Good
2014	Hwang *et al.*	53/F	Comatose	Distal LSA	ICH (BG) + IVH	Embolization with the LSA sacrifice	Good
		44/F	Comatose	Distal LSA	ICH (BG) + IVH	Embolization with the LSA sacrifice	Good
2016	Liu *et al.*	10/M	?	Distal LSA	IVH	Bilateral EDAS → disappeared	Good
2018	Ni *et al.*	37/F	Comatose	Distal LSA	ICH (BG) + IVH	STA-MCA → disappeared	Good
2022	Byeon *et al.*	42/M	Headache, nausea	Distal LSA	ICH + IVH	Embolization with the LSA sacrifice	Good
2022	Fu *et al.*	73/M	Headache, nausea, vomiting	Distal LSA (RAH)	ICH (BG)	Embolization failed → conservation	Poor (died)
2023	Ando *et al.*	42/F	Right hemiparesis, motor aphasia, facial palsy	Distal LSA	ICH (BG)	Embolization without the LSA sacrifice	Good
2023	Zhou *et al.*	56/M	?	Distal LSA	IVH	Embolization with the LSA sacrifice	Good
		40/F	?	Distal LSA	IVH	Embolization with the LSA sacrifice	Good
		65/F	?	Distal LSA	ICH	Embolization with the LSA sacrifice	Good
2024	Hashio *et al.*	29/F	?	Distal LSA	ICH (BG) + SAH	surgical hematoma removal + decompressive craniectomy → rebleeding → surgical excision	Good
2024	Present case	56/F	Headache, nausea, vomiting	Distal LSA	ICH (BG) + IVH	Embolization with the LSA sacrifice	Good

Abbreviations: BG, basal ganglia; EDAS, encephalo-duro-arterio synangiosis; ICH, intracerebral hemorrhage; IVH, intraventricular hemorrhage; LSA, lenticulostriate artery; MMD, moyamoya disease; RAH, recurrent artery of Heubner; SAH, subarachnoid hemorrhage; STA-MCA, superficial temporal artery-middle cerebral artery.
* The RAH is the largest of the medial LSAs.

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